

Case reports

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Double outlet right ventricle with spontaneously developing pulmonary outflow obstruction

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A case of double outlet right ventricle Type II A developing obstruction of the outflow from the right ventricle to the pulmonary artery is described. Life-long follow-up and re-evaluation of operability are indicated in similar cases.

Double outlet right ventricle is a well-defined and often correctable type of congenital heart disease. The disease has been divided into two groups according to the position of the ventricular septal defect (Neufeld *et al.*, 1962). In Type I the defect

is situated posteroinferiorly to the crista supraventricularis and in Type II the defect lies anterosuperiorly to the crista supraventricularis. Type II can be divided into two subgroups: II A with the defect close to the pulmonary valve and II B with

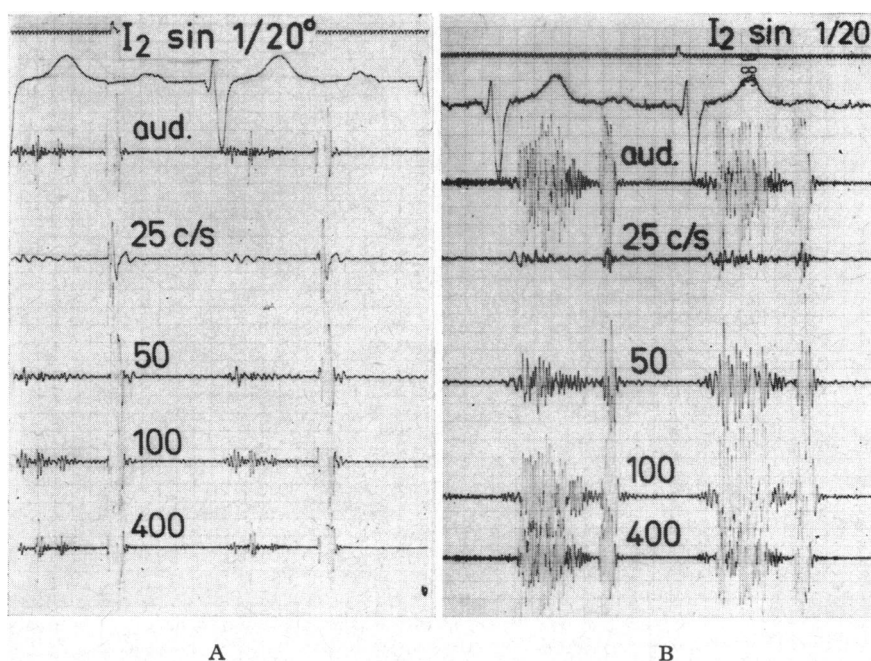


FIG. 1 A and B. Phonocardiogram recorded from the 2nd left interspace at the age of 15 years (Fig. 1 B) shows a strong high-frequency systolic murmur which is not present at the age of 12 years (Fig. 1 A).

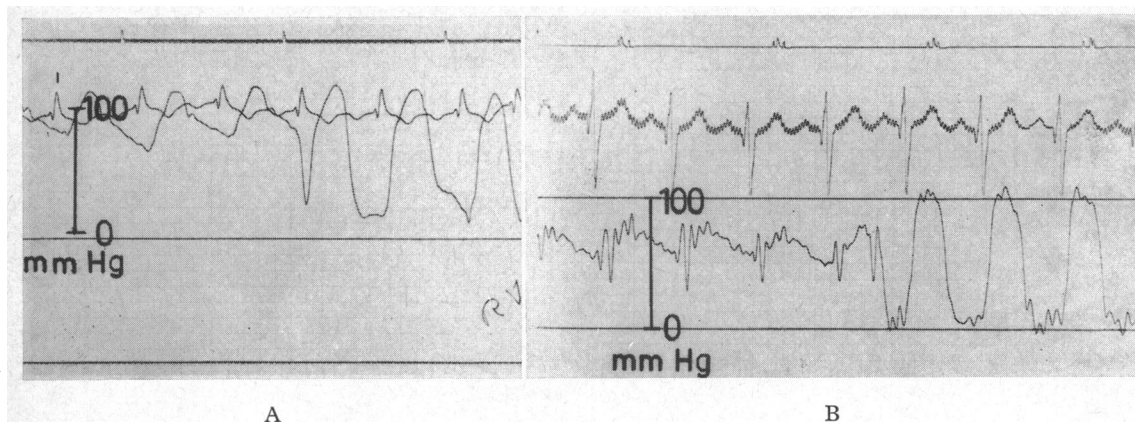


FIG. 2 A and B. The withdrawal curve from the pulmonary artery to the right ventricle at the age of 15 years (Fig. 2 B) showing a pressure gradient of 25 mmHg; the gradient is absent at the age of 12 years (Fig. 2 A).

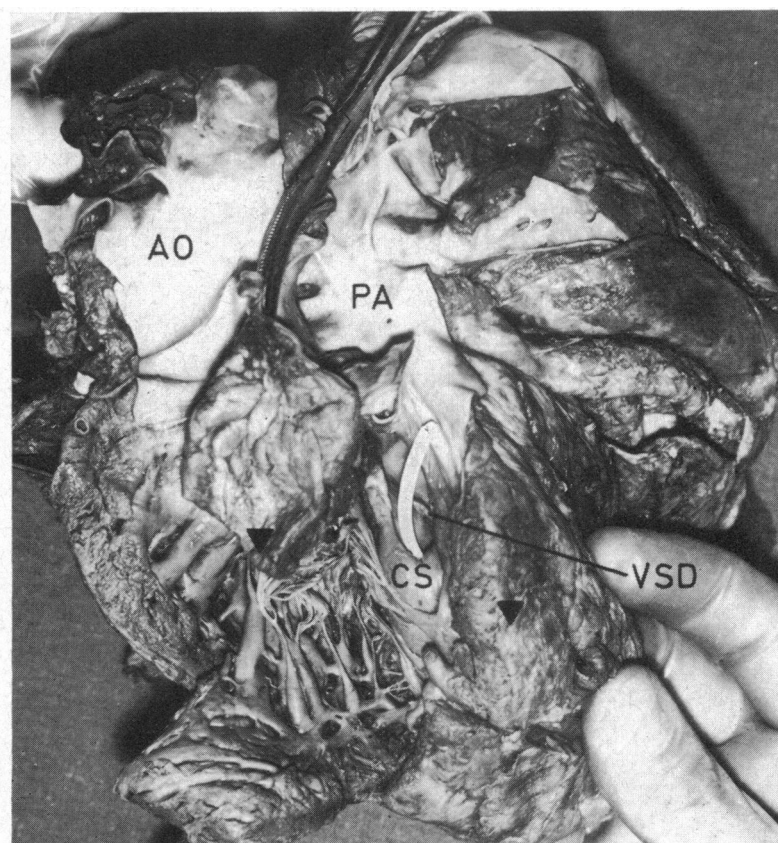


FIG. 3 The right ventricle with the aorta (AO) and the pulmonary artery (PA) side by side. The white probe is passed through the ventricular septal defect (VSD) from the left ventricle to the pulmonary artery. The defect is situated above the crista supraventricularis (CS). The parietal limb of the crista supraventricularis is divided and the cut surfaces are marked by filled triangles (cf. Fig. 4).

the defect below the pulmonary and the aortic valves. Double outlet right ventricle may be associated with pulmonary stenosis (Dayem *et al.*, 1967; Neufeld, DuShane, and Edwards, 1961; Witham, 1957; Hallermann *et al.*, 1970; Shetty and Martin, 1967). According to Edwards *et al.* (1965) this association is found only in cases with infracristal defects (Type I). In the published reports we have found only one case acquiring infundibular obstruction (Engle *et al.*, 1963; Redo *et al.*, 1963): this case, which was of Type I, had been irradiated for a neuroblastoma of the mediastinum. The patient to be described, who developed pulmonary outflow obstruction, is apparently of Type II A.

Case report

The patient, a boy, was born in 1955. His birthweight was 3.75 kg. Cardiac failure with severe cyanosis was noted from the age of 2 weeks. At 1 year he weighed 4 kg. From his second year of life he improved and gained weight, but his physical activity was restricted to a minimum. At the age of 12 years, a weak (grade 1/6–2/6), early systolic murmur was heard at the 2nd left intercostal space (Fig. 1 A). This murmur had been present since infancy. Cyanosis and clubbing of his fingers were extreme. The haematocrit was 80 per cent. Cardiac catheterization and angiocardiology performed in November 1967 verified the diagnosis of double outlet right ventricle. The peak systolic pressures in the right ventricle, the pulmonary artery, and the aorta were

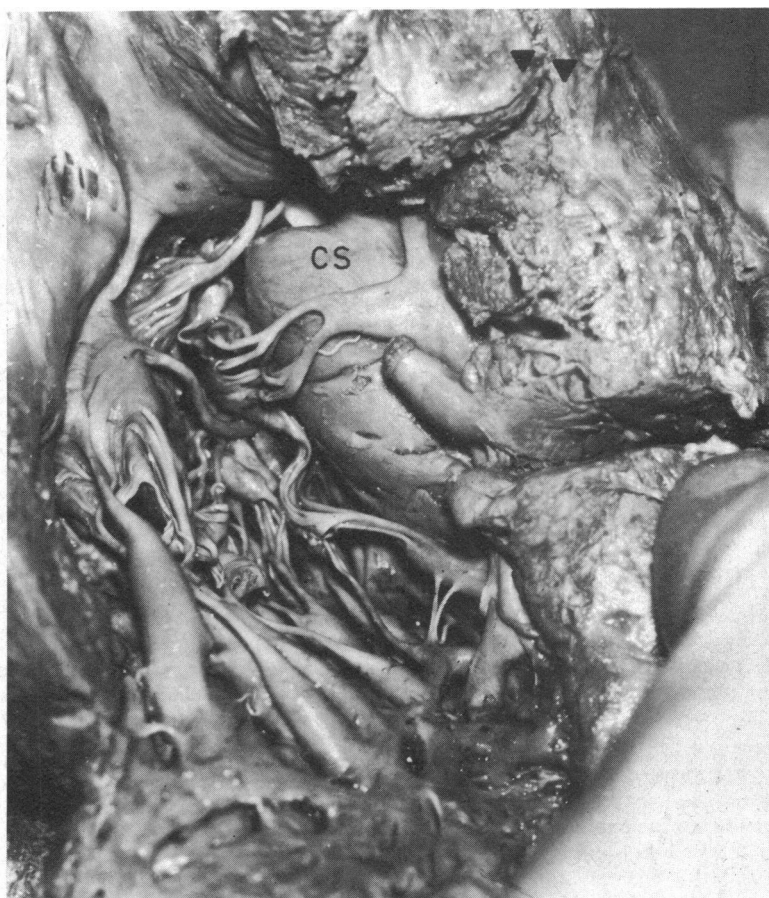


FIG. 4 Pulmonary outflow tract of the right ventricle. The cut surfaces of the parietal limb of the crista supraventricularis, marked by filled triangles, have been approximated to show the obstruction of the outflow. The white probe from the left ventricle through the ventricular septal defect is visible above the crista supraventricularis.

105 mmHg. The oxygen saturation in the aorta was 62 per cent and in the pulmonary artery 78 per cent. There was no pressure gradient between the pulmonary artery and the right ventricle (Fig. 2 A). Between his thirteenth and his sixteenth year a systolic thrill was noticed and a harsh grade 4-5/6 early midsystolic murmur was heard at the second left intercostal space (Fig. 1 B). He was extremely cyanotic and his haematocrit was 87 per cent. Further catheterization was performed in December 1970. The systolic right ventricular pressure was 110 mmHg, the pulmonary artery pressure 85 mmHg, the peak systolic gradient at rest being 25 mmHg (Fig. 2 B). The saturation in the pulmonary artery was 80 per cent. An atrial septectomy as described by Blalock and Hanlon was made. After the operation his physical capacity was greatly improved and his cyanosis decreased. The systolic murmur did not change. The haematocrit was stabilized at 63 per cent. As his general condition improved, he started to ride a motorcycle. He died in a motor accident about a year after the operation.

Necropsy

The heart was enlarged predominantly from the hypertrophy of the right ventricle. The right ventricular wall was 13 mm and the left ventricular wall was 8 mm thick. The crista supraventricularis was greatly hypertrophied thus obstructing the outflow from the right ventricle to the pulmonary artery (Fig. 3 and 4). The area at the narrowest part was estimated to be 0.3 cm². The left ventricular cavity was of normal size with a normal mitral valve. The only exit was through the ventricular septal defect. The aorta and the pulmonary trunk originated separately from the right ventricle, the aorta to the right of the pulmonary trunk. The ventricular septal defect was situated antero-superiorly to the crista supraventricularis, partly adjacent to the pulmonary valve. The size of the defect was 28 × 15 mm. One of the three cusps of the pulmonary valve was somewhat thickened; the other two were normally mobile. The area of the valve was estimated to be 1 cm².

Discussion

As judged by the site of the ventricular septal defect, the case here described belongs to Type II A. The clinical course with the appearance of the intense and harsh murmur and the pressure gradient supports the diagnosis of acquired pulmonary outflow obstruction which later was verified at necropsy. The Type II deformity is supposed not to be combined with infundibular pulmonary stenosis. In this case there was no stenosis when the patient was examined in 1967, but in 1970 a pressure difference between

the right ventricle and the pulmonary trunk was evident. The patient had no history of endocarditis during this time. Obstruction of the pulmonary outflow tract was found at necropsy.

In Type II A most of the left ventricular blood enters the pulmonary trunk, while the aorta receives blood predominantly from the right ventricle. This explains the improvement obtained by venous admixture. This patient with longstanding pulmonary hypertension was judged to be inoperable. The occurrence of a progressive infundibular pulmonary outflow obstruction in similar cases could make correction possible, at least in children where the pulmonary hypertension has been of shorter duration. There is great variability in the anatomy and the natural history of double outlet right ventricle, as exemplified by this case. Thus, follow-up and re-evaluation are necessary.

References

- Dayem, M. K. A., Preger, L., Goodwin, J. F., and Steiner, R. E. (1967). Double outlet right ventricle with pulmonary stenosis. *British Heart Journal*, **29**, 64.
- Edwards, J. E., Carey, L. S., Neufeld, H. N., and Lester, R. G. (1965). Origin of both great vessels from the right ventricle with pulmonary stenosis. In *Congenital Heart Disease*, Vol. II, p. 486. Saunders, Philadelphia.
- Engle, M. A., Steinberg, I., Lukas, D. S., and Goldberg, H. P. (1963). Acyanotic ventricular septal defect with both great vessels from the right ventricle. *American Heart Journal*, **66**, 755.
- Hallermann, F. J., Kincaid, O. W., Ritter, D. G., Ongley, P. A., and Titus, J. L. (1970). Angiocardiographic and anatomic findings in origin of both great arteries from the right ventricle. *American Journal of Roentgenology*, **109**, 51.
- Neufeld, H. N., DuShane, J. W., and Edwards, J. E. (1961). Origin of both great vessels from the right ventricle. *Circulation*, **23**, 603.
- Neufeld, H. N., Lucas, R. V., Jr., Lester, R. G., Adams, P., Jr., Anderson, R. C., and Edwards, J. E. (1962). Origin of both great vessels from the right ventricle without pulmonary stenosis. *British Heart Journal*, **24**, 393.
- Redo, S. F., Engle, M. A., Holswade, G. R., and Goldberg, H. P. (1963). Operative correction of ventricular septal defect with origin of both great vessels from the right ventricle. *Journal of Thoracic and Cardiovascular Surgery*, **45**, 526.
- Shetty, A. V., and Martin, R. (1967). Double outlet right ventricle with pulmonary stenosis. *British Heart Journal*, **29**, 279.
- Witham, A. C. (1957). Double outlet right ventricle. *American Heart Journal*, **53**, 928.

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